## PRODUCT INFORMATION

## Expression system

E.coli

Domain
1-97aa
UniProt No.
060220

## NCBI Accession No.

NP_004076

## Alternative Names

Mitochondrial import inner membrane translocase subunit Tim8 A, Mitochondrial import inner membrane translocase subunit Tim8 A, DDP, DDP1, DFN1, MTS, TIM8

## PRODUCT SPECIFICATION

## Molecular Weight

13.4 kDa (120aa) confirmed by MALDI-TOF

## Concentration

$0.25 \mathrm{mg} / \mathrm{ml}$ (determined by Bradford assay)

## Formulation

Liquid in. 20 mM Tris-HCl buffer (pH 8.0) containing $0.15 \mathrm{M} \mathrm{NaCl}, 30 \%$ glycerol, 1 mM DTT

## Purity

> 90\% by SDS-PAGE

## Tag

His-Tag

## Application

SDS-PAGE

## Storage Condition

Can be stored at +2 C to +8 C for 1 week. For long term storage, aliquot and store at -20 C to -80C. Avoid repeated freezing and thawing cycles.

## BACKGROUND

## Description

TIMM8A is involved in the import and insertion of hydrophobic membrane proteins from the cytoplasm into the mitochondrial inner membrane. The gene is mutated in Mohr-Tranebjaerg syndrome/Deafness Dystonia Syndrome (MTS/DDS) and it is postulated that MTS/DDS is a mitochondrial disease caused by a defective mitochondrial protein import system. Defects in this gene also cause Jensen syndrome; an X-linked disease with opticoacoustic nerve atrophy and muscle weakness. This protein, along with TIMM13, forms a 70 kDa

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## Recombinant human TIMM8A protein

Catalog Number: ATGP2486
heterohexamer. Alternative splicing results in multiple transcript variants encoding distinct isoforms. Recombinant human TIMM8A proten, fused to His-tag at N-terminus, was expressed in E. coli and purified by using conventional chromatography techniques.

## Amino acid Sequence

MGSSHHHHHH SSGLVPRGSH MGSMDSSSSS SAAGLGAVDP QLQHFIEVET QKQRFQQLVH QMTELCWEKC MDKPGPKLDS RAEACFVNCV ERFIDTSQFI LNRLEQTQKS KPVFSESLSD

## General References

Rothbauer u., et al. (2001) J. Biol. Chem. 276:37327-37334
Roesch K., et al. (2004) Hum. Mol. Genet. 13:2101-2111

DATA


